Cavernous sinus syndrome: a case report

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Abstract

Background: Cavernous sinus syndrome (CSS) is a rare, but potentially life- and sight-threatening condition. We report a case of multiple cranial neuropathies with CSS.

Case presentation: A 70-year-old woman presented with unilateral complete ptosis, ophthalmoplegia, and diplopia. Examination showed left-sided multiple cranial nerve palsies, with the involvement of cranial nerve III (oculomotor nerve), IV (trochlear nerve), V1 and V2 (ophthalmic and maxillary branches of trigeminal nerve), and VI (abducens nerve). A clinical diagnosis of CSS was made. Neuroimaging revealed a left-sided cavernous sinus mass for further investigation.

Conclusion: In this case report, we highlight the clinical and radiological features of CSS to raise clinical suspicion for similar diagnosis in the future.

Keywords: cavernous sinus syndrome, cranial nerve palsies, ophthalmoplegia

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Sindrom sinus cavernous: laporan kes

Abstrak

Latar belakang: Sindrom sinus cavernous (CSS) adalah jarang terjadi tetapi ianya berpotensi menyebabkan kehilangan nyawa dan penglihatan. Kami melaporkan kes (CSS) yang melibatkan neuropati kepada beberapa saraf kranial.

Laporan kes: Seorang wanita berusia 70 tahun mengalami ptosis yang teruk, oftalmoplegia dan diplopia. Hasil dari pemeriksaan klinikal terdapat penglibatan saraf kranial sebelah kiri di mana termasuk saraf kranial III (saraf okulomotor), IV (saraf troklear), V1 dan V2 (cabang oftalmik dan maxillari daripada saraf trigeminal) dan VI (saraf abdusen). Pengimejan neurologi telah dilakukan dan didapati terdapat ketumbuhan pada sinus cavernous sebelah kiri yang memerlukan siasatan lanjut.

Kesimpulan: Dari laporan kes ini, bagi meningkatkan pengesanan sindrom CSS, penekanan ke atas ciri-ciri klinikal dan radiologi adalah penting.

Keywords: sindrom sinus cavernous, palsi saraf kranial, oftalmoplegia

Introduction

Cavernous sinus syndrome (CSS) is a rare but potentially life and sight-threatening condition, by which the suspicion for diagnosis should be raised on clinical grounds. CSS presents as multiple cranial neuropathies with variable involvement of cranial nerves (CN) III, IV, V1, V2, and VI with or without additional signs and symptoms such as proptosis, conjunctival injection, retroorbital pain, and headache. Symptoms may be acute or progressive depending on the underlying aetiology. In some cases, clinical manifestation of CSS is non-specific with variable combinations of symptoms, further complicating the diagnostic process. A good understanding of the anatomical structures traversing the cavernous sinus, the pathophysiology of the disease process and the clinical manifestation of the disease can greatly assist in making a prompt diagnosis to improve prognosis.

Case presentation

A 70-year-old woman with underlying diabetes mellitus, hypertension, dyslipidaemia, and history of right occipital lobe infarction presented with left droopy eyelid and double vision for 2 weeks. She also complained of progressive blurring of vision in her left eye associated with intermittent headache. There were no symptoms of
increased intracranial pressure or other neurological symptoms. No prior history of trauma preceded the event.

On examination, her best-corrected visual acuity was 6/18 in the right eye and 6/120 in the left eye. The left eye showed complete ptosis and 15° exotropia on primary gaze with dilated pupil (Fig. 1 a, b). There was complete reduction of extraocular movement on all gaze directions with power of 1/5 remaining on left lateral gaze (Fig. 1c). There was no relative afferent pupillary defect. Conjunctiva was white with no dilated tortuous episcleral vessels. Corneal reflex was reduced in the left eye, with ipsilateral hypoesthesia over facial territories supplied by the ophthalmic and maxillary branches of the left trigeminal nerve. Otherwise, both the anterior and posterior segments of the left and right eyes were normal. The patient was clinically diagnosed with CSS.

Fig. 1. Ocular manifestations of cavernous sinus syndrome in primary gaze. (a) Complete left eye ptosis. (b) Left eye exotropia and hypotropia. (c) Extraocular movement in nine cardinal positions of gaze. Presence of left eye ophthalmoplegia in all gaze directions.
An urgent contrasted computerized tomography (CT) of the brain and orbit was performed. There was a heterogeneously enhancing lesion present in the left cavernous sinus with anterior extension to the orbital apex and left Meckel's cave. There were also radiological features of multifocal infarction, cerebral atrophy, and small vessel disease over the right occipital lobe, bilateral basal ganglia, right corona radiata, and centrum semiovale. Further magnetic resonance imaging (MRI) of the brain showed soft tissue thickening in the left cavernous sinus (Fig. 2). Differential diagnosis at this point included inflammatory changes, infectious causes, and tumour infiltration. Adjunctive tests performed included a complete blood count with differential counts, erythrocyte sedimentation rate, C-reactive protein, complete metabolic panel, tumour markers, intradermal tuberculin test, sputum for acid fast bacilli, syphilis, and chest X-ray. The tests were unremarkable. Our patient was co-managed with the neurosurgical and otorhinolaryngology teams following the imaging findings. A nasal endoscopy performed by the otorhinolaryngology team showed a right torus tuleres mass and a mildly bulging left fossa of Rosenmuller. Biopsy of the nasal mass showed enlarged lymphoid follicles with no trace of malignancy. Patient was counselled for further interventions, which included a lumbar puncture and a biopsy of the intracavernous lesion to guide further management. However, our patient refused further investigation in view of her old age and personal logistic issues. Follow-up to her condition was discontinued due to the patient's death to COVID 3 months following the diagnosis.

Discussion

The cavernous sinus is a small yet complex and important structure, which comprises the dural venous sinus located at the central base of skull on either side of the sella turcica containing the pituitary gland. It houses the CN III, IV, V1, V2, and VI, as well as the internal carotid artery and sympathetic fibres. CSS is defined as the involvement of at least two of the intracavernous CNs, or involvement of only
Cavernous sinus syndrome: a case report

one CN in combination with radiologically confirmed cavernous sinus lesions. It is caused by pathological lesions within the cavernous sinus leading to compression and compromise of the intracavernous neurovascular structures located within the small venous space. The true incidence of CSS is not well documented, with few studies conducted regarding its epidemiology. Approximately 5% of ophthalmoplegia in the United States were reported to be secondary to the CSS. CSS affects both genders equally.

Diplopia and headache are the most common presenting symptoms reported among patients diagnosed with CSS, presenting in up to 90% of patients with CSS. Other common presenting symptoms and signs reported include retro-orbital pain, facial paraesthesia, vision loss, hearing loss, proptosis, ptosis, painful or painless ophthalmoplegia, decreased corneal reflexes, and seizures. Physical examination findings of CSS are dependent on CN involved, with the most common CN involved being CN III (85%), followed by CN VI (70%), V1, V2, and IV. The involvement of all CNs in CSS, as was evident in our case, is in fact uncommon. A study conducted by Fernández et al. in Spain showed that complete ophthalmoplegia was present in only 17% out of a series of 126 patients. Painful ophthalmoplegia is less common and has been reported to be associated with inflammatory causes. The involvement of ophthalmic and maxillary branches of the trigeminal nerve embedded in the lateral walls of the cavernous sinus explains the ipsilateral upper facial hypoaesthesia, sparing the lower facial dermatome, which is supplied by V3 branch. The involvement of pupil dilatation may or may not be present in CSS due to the concurrent presence of both sympathetic and parasympathetic innervations in the cavernous sinus. In view of the direct venous drainage of the cavernous sinus from several facial structures including the eye, large cavernous sinus lesions may also exert mass effect leading to less commonly reported signs such as proptosis, conjunctival congestion, and ocular hypertension. These signs were not evident in our patient.

CSS can be caused by a broad category of diseases. Tumours account for the majority of CSS cases. These neoplastic lesions can be primary or metastatic. Primary neoplastic lesions include meningioma, schwannoma of cranial nerves in the cavernous sinus, neurogenic tumour, and haemangioma. Metastatic lesions can arise from the breast, lung, and prostate, or via local spreading from facial structures, such as nasopharyngeal carcinoma with intracranial and cavernous sinus extension, which is highly prevalent in Southeast Asian countries and seemingly the culprit in our patient. Other causes of CSS include trauma, vascular pathologies (i.e., carotid-cavernous fistula, carotid-cavernous aneurysm, or cavernous sinus thrombosis), infections (i.e., tuberculosis, mucormycosis, or Aspergillosis) and inflammation (i.e., Tolosa-Hunt syndrome or Wegener granulomatosis). The wide variety of possible aetiologies contributes to the diagnostic challenge. Hence, the involvement of multidisciplinary teams in the diagnostic and management process is required to optimize the patient’s outcome.
Neuroimaging plays a huge role in establishing a definitive diagnosis of CSS. Contrasted CT scan is useful in providing visualization of the cranial bones and the adjacent structures. MRI, on the contrary, is the most sensitive tool for visualizing soft tissues.¹ Both serve as valuable evaluation tools in the identification of causative lesion of CSS and its anatomical relations, as well as to identify the extent of local spread and source of metastasis in cases of tumours.⁴ CT angiography and MR angiography can be acquired for further investigation of CSS of vascular aetiologies. Adjunctive investigation tools such as lumbar puncture and lesion biopsy are invasive and should be reserved for cases where diagnostic capability is limited based on clinical and imaging findings alone. In fact, some studies found lumbar puncture to be a poor diagnostic tool with low sensitivity in CSS cases.¹ Ancillary blood tests to rule out infectious and inflammatory causes are warranted in cases where common causes such as tumours and vascular aetiology have been excluded.

**Conclusion**

CSS presents as multiple CN III, IV, V1, V2, and VI palsies with variable combinations of symptoms. The diagnostic process can be challenging, yet crucial, given the life- and sight-threatening nature of the disease. Understanding the characteristic clinical features of CSS can assist in differential diagnosis. An early neuroimaging study is recommended in all patients with CSS. Prompt diagnosis of CSS and its underlying aetiology can improve prognosis.

**Declarations**

**Consent for publication**
The patient provided informed consent for the publication of this case report.

**Competing interests**
None to declare.

**Funding**
None to declare.

**Acknowledgements**
None to declare.
References